

Importance and implications of the occurrence of supraventricular arrhythmia in children with Catecholaminergic Polymorphic Ventricular Tachycardia.

*Shkolnikova M., Bereznitskaya V., Termosesov S.
Moscow Institute for Pediatrics and Pediatric Surgery*

Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) in children is rare malignant arrhythmia with a high risk of syncope and sudden cardiac death (SCD) due to the stress-induced polymorphic VT. In addition to ventricular arrhythmias different types of supraventricular tachyarrhythmia (SVT) is the common cause of inappropriate ICD shocks. The aim was to assess the prevalence and clinical significance of SVT types in children with CPVT.

Patients and Methods. 24 pts (15 males -63%) aged from 4.5 to 14.0 with exercise- induced CPVT and syncope were enrolled in the study. Family and clinical history, 12-leads ECG, 24-hour ECG monitoring, stress-testing, echocardiography, blood chemistry, end EP study (11 pts) were done. Follow-up time varied from 3 to 18 years.

Results. According to Kaplan-Meier survival analysis, 50% of CPVT children experienced first syncope before the age of 5 and more than 35% developed aborted cardiac death or SCD before the age of 13. Family history of SCD or syncope took place in 38% of pts. ECG and Holter revealed sinus bradycardia in 83%, PQ interval \leq 110 ms – in 68%, ventricular premature beats –in 55%, and AV-dissociation – in 55% of pts. SVT as a trigger of CPVT was confirmed in 77% of pts. Among them typical or atypical atrial flutter (AFI) was found in 32% of cases. 11 pts were implanted with ICD. Inappropriate shocks were mostly associated with SVT with high ventricular response. Adding to the beta-blocker therapy the sodium-channel blockers has allowed to control supraventricular triggers and improve the prognosis. In one patient with polytopic SVT and high AV conduction (210 bpm) AV node cryomodification reduced ventricular rate to 125 bpm.

Conclusions. Vulnerability to ventricular and also to supraventricular arrhythmia characterizes young patients with CPVT. SVT including atrial flutter is significant as trigger of malignant ventricular arrhythmia and as a cause of inappropriate ICD shocks. Combined antiarrhythmic therapy and cryomodification of AV-node could be suggested for SV triggers control.